A rare case of lung tumor - pulmonary inflammatory pseudotumor

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Abstract

Pulmonary inflammatory pseudotumor (PIP) is a rare condition of unknown etiology. It is still a matter of debate if it represents an inflammatory lesion characterized by uncontrolled cell growth or a true neoplasm[1,2]. Although mostly benign, these tumors are diagnosis and therapeutic challenges. Preoperative diagnosis can rarely be established. The treatment of choice is surgical resection which has both diagnostic and therapeutic value. We report the case of a 63-year-old male presented with clinical and imagistic picture suggestive of malignancy in the thorax. Lobectomy was performed with histological diagnosis of PIP. No evidence of tumor recurrence.
Keywords: *inflammatory pseudotumor, lung, malignancy*

Rezumat

Un caz rar de tumoră pulmonară – pseudotumoră inflamatorie pulmonară
Pseudotumora inflamatorie pulmonară (PIP) este o entitate rară, de cauză necunoscută. Încă se dezbate dacă este o leziune inflamatorie caracterizată prin creştere celulară necontrolată sau un neoplasm adevărat[1,2]. Deşi de cele mai multe ori de natură benignă, aceste tumori reprezintă provocări de diagnostic şi tratament. Diagnosticul preoperator de certitudine poate fi rareori stabilit. Tratamentul de elecţie este rezecţia chirurgicală care are valoare atât diagnostică, cât şi terapeutică. Prezentăm cazul unui pacient de 63 de ani, care s-a prezentat cu un tablou clinic şi imagistic sugestiv de tumoră pleurală. S-a practicat o lobectomie cu diagnosticul histologic de PIP. Nu a existat recidivă tumorală postoperatorie.
Cuvinte-cheie: *pseudotumoră inflamatorie, plămân, malignitate*

Background

Pulmonary inflammatory pseudotumor (PIP) is a rare condition of unknown etiology. It was first described in the lung by Brunn in 1939 and Umiker gave its name in 1954, due to clinically and radiological similarity to a malignant process[3]. Inflammatory pseudotumor most commonly involves the lung and the orbit, but the studies showed that it can occur almost anywhere in the body (stomach, kidney, central nervous system, gastrointestinal tract etc.). This condition has been accepted as a benign process for a long period of time, consisting of aberrant or exaggerated response to tissue injury undergoing a chronic inflammatory process, despite the fact that sometimes the features are those of a spindle cell neoplasm. Although usually it has a benign evolution, a small number of cases develop local recurrence, infiltrative local growth with the development of multifocal, noncontiguous tumors, vascular invasion, and malignant transformation[4].

Case presentation

We present the case of a 63-year-old male with a history of 52 pack years of cigarette smoking. His past medical history comprise of: ischaemic heart disease and systemic hypertension since 2002, pulmonary tuberculosis in 2005 (treated and cured), diabetes mellitus since 2007, peripheral obliterative arteriopathy, and COPD (GOLD stage II) diagnosed in 2004, with a previous COPD exacerbation in March 2012, when he received antibiotics and systemic corticosteroids with good outcome. He was admitted in our department in September 2012 with a 3-month history of mild hemoptysis. Physical examination was normal (none of the following: fever, peripheral lymphadenopathy, peripheral edema, pulmonary rales; the blood pressure was 140/80 mmHg, the pulse was 73 beats/minute, the respiratory rate was 16 breaths/minute, and the oxygen saturation was 96% while breathing ambient air). Results of laboratory tests were normal. Pulmonary function tests identified a moderate obstructive syndrome. Post-bronchodilator spirometry values were: FEV1/FVC ratio 0.54, FVC 2.84L (71.2% predicted), FEV1 1.60L (51.2% predicted). Postero-anterior Chest X-Ray showed only hyperinflation (Figure 1).

Due to his symptomatology (persistent mild hemoptysis) and the heavy history of smoking with a high risk of lung cancer, we performed a high-resolution computed tomography

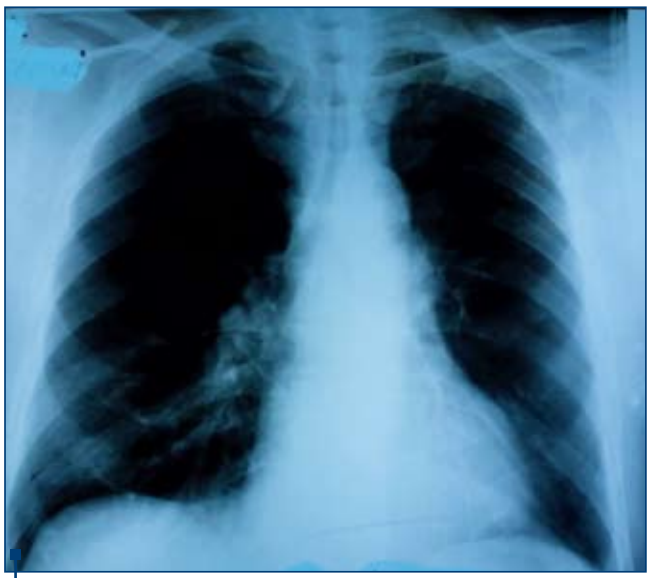

Figure 1. *Postero-anterior chest radiography showing hyperinflation*

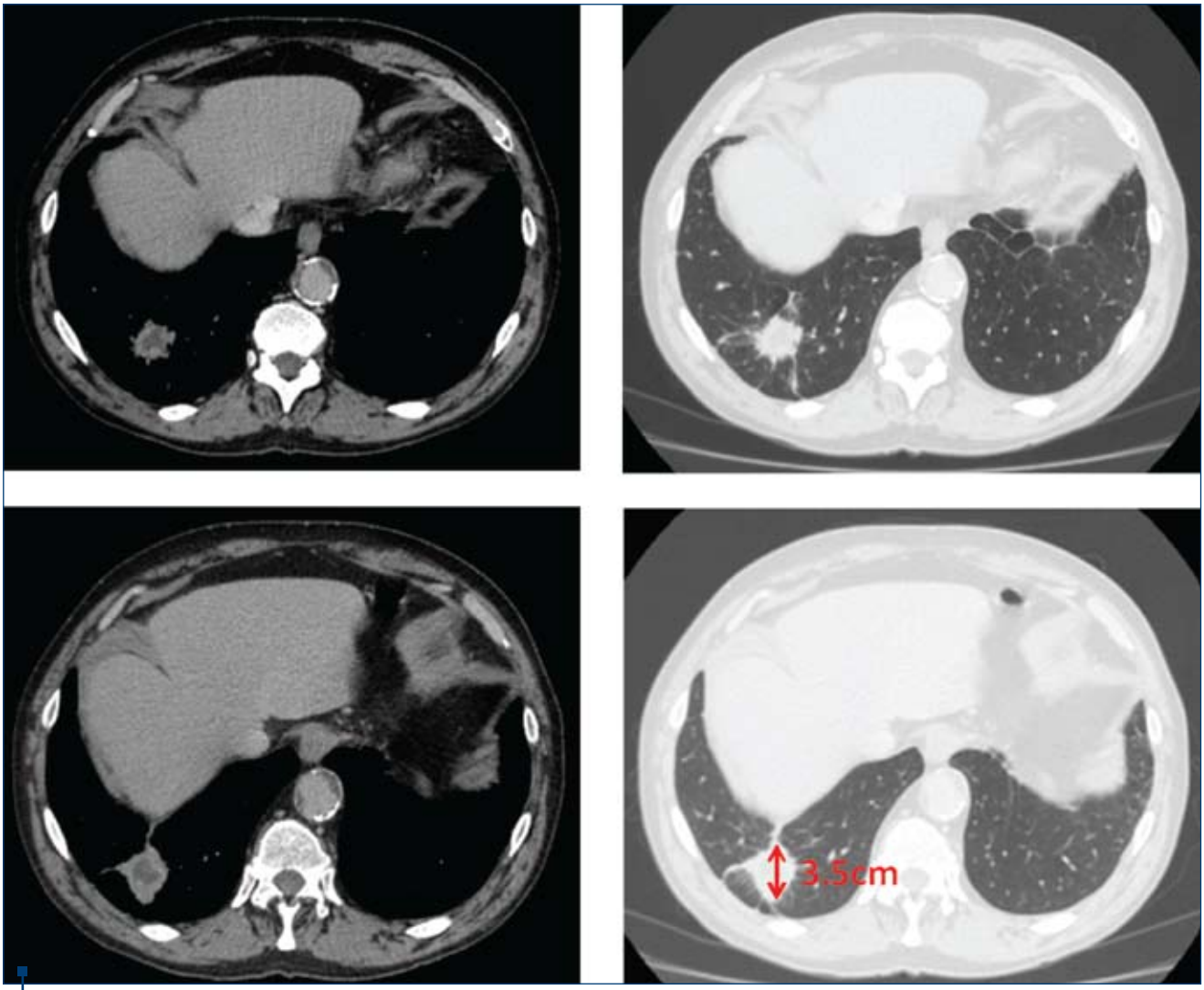

Figure 2. High-resolution computed tomography showing a heterogeneous pulmonary tumor mass, with spicular radiation, in the right lower lobe

with contrast that showed a tumor mass of 3.5 cm, round, with spicular radiation, with an area of central lucency located in right lower lobe (Figure 2), without pleural indentation. No associated pleural effusion, mediastinal lymphadenopathy or other opacities. This aspect was highly suggestive of lung cancer. Sputum bacterial cultures were negative. Smears for acid fast bacilli were negative. Bronchoscopy showed no bronchial tumor. Bronchoalveolar lavage showed only an elevated total cell number (15x10^6cells/100mL), with mild increase of neutrophils (5%). It was also negative for acid-fast bacilli and tumor cells.

By that moment it was highly suggestive of lung cancer, so we decided to send the patient to thoracic surgery. We performed several other investigations to ensure that this patient was operable. Complex pulmonary function tests showed: normal total lung capacity (7.39L, 113.7% predicted), hyperinflation with high residual volume (4.82L, 202.1% predicted) and a moderate reduction of the transfer factor (45.7% predicted). On 6 minute walk test patient walked 490 m, with a significant oxygen desaturation from 94% to 89% while breathing ambient air. Cardiac ultrasonography revealed concentric left ventricular hypertrophy, hypokinesis of interventricular septum in basal region, with normal ejection fraction. We decided along with the thoracic surgeons that the patient was operable, and right inferior lobectomy was performed. Histopathology report showed no evidence of malignancy, with the presence of heavy inflammatory cell infiltrate composed predominantly of lymphocytes and plasma cells. It was also noted focal areas of micro-abscess formation with necrosis and a marked degree of fibrosis (inflammatory pseudotumor). The patient recovered slowly after the intervention. Five months after surgical resection patient showed no local recurrence.

Discussions

Studies report inflammatory pseudotumor to comprise 0,04 to 1% of all lung tumors[5, 6], but there are the most common benign lung tumors in children under 16 years of age[5, 6]. Although inflammatory pseudotumor can develop at any age, it is mostly a disease of young adults. There is no sex predilection. Even inflammatory pseudotumors are generally regarded as inflammatory lesions, sometimes they may have anaplastic features, as local invasion or recurrence, distant

metastases, and cytogenetic clonal changes[7]. The etiology and pathogenesis remains uncertain. Like in the case of our patient, a history of prior infection was demonstrated in one third of the patients[8]. Histopathologically, the aspect comprise of a variable mixture of collagen, inflammatory cells and benign mesenchymal cells. As consequence, there is a variable terminology to describe PIP, like the following: plasma cell granuloma, histiocytoma, fibroxanthoma, inflammatory myofibroblastic tumor, xanthoma and xanthogranuloma, and plasma cell-histiocytoma complex. Some authors believe this tumor is a low-grade fibrosarcoma with inflammatory cells. Only a small number of patients with PIP are symptomatic. The usual presenting symptoms are cough, fever, dyspnea, and hemoptysis. The laboratory data are no conclusive. The radiological features of PIP usually shows a solitary peripheral nodule or mass, typically peripheral and in the lower lobes[9]. Occasionally, patients may present areas of pneumonic consolidation or atelectasis, endobronchial lesions, multiple nodular lesions, hilar lymphadenopathy or pleural involvement. Radiographic images, sputum culture and sputum cytology, bronchoscopy with bronchoalveolar lavage and percutaneous fine needle aspiration biopsy are considered insufficient for diagnosis. Surgical resection is the treatment of choice because surgery makes both the correct diagnosis and complete cure. When there is delay in diagnosis and treatment of PIP, the lesion may evolve mainly with local invasion. The surgical approach is the complete resection due to the risk of recurrence. Local recurrence is due to incomplete resection of the primary lesion. Metastasis of the tumor has been described[10] and the local recurrence rate after complete resection is low[10].

Conclusions

Pulmonary inflammatory pseudotumor is a rare entity, but the resemblance with the lung cancer is inducing the same diagnosis and therapeutic approach in both cases. The treatment of choice is surgical and complete resection leads to an excellent prognosis. Long term follow-up is needed. ■

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